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The role of nursing staff in the nutritional therapy of a child with cystic fibrosis

Rola personelu pielęgniarskiego w terapii żywieniowej dziecka z mukowiscydoza

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Abstract

Cystic fibrosis (CF) is classified as a rare genetic disease and at the same time is the most common inherited disease in an autosomal recessive way. In Poland the disease is diagnosed in 1 in 4394-5000 live neonatal births [7, 9]. CF is a systemic chronic disease with progressive progression, incurable [10]. The cause of most problems in patients with CF are: chronic obstructive lesions and respiratory tract infections and impaired digestion processes with their consequences. The basis in the patient care of CF is the education of the patient/his caregivers in the prevention and treatment of bronchopulmonary disease and modified nutrition rules.

Streszczenie

Mukowiscydoza (CF) jest zaliczana do rzadkich chorób genetycznych, a jednocześnie jest najczęstszą chorobą dziedziczoną w sposób autosomalny recesywny. W Polsce chorobę rozpoznaje się u 1 na 4394-5000 żywo urodzonych noworodków [7, 9]. CF jest ogólnoustrojową chorobą przewlekłą, o postępującym przebiegu, nieuleczalną [10]. Przyczyną większości problemów u pacjentów z CF, są: przewlekłe zmiany obturacyjne i infekcje dróg oddechowych oraz zaburzenia procesów trawienia wraz z ich konsekwencjami. Podstawą w opiece nad pacjentem z CF jest edukacja pacjenta/ jego opiekunów w zakresie postępowania profilaktyczno-leczniczego choroby oskrzelowo-płucnej i zmodyfikowanych zasad żywienia pacjenta.

Key words: cystic fibrosis, nutrition

Słowa kluczowe: mukowiscydoza, żywienie

Cystic fibrosis CF – Cystic fibrosis, mucoviscidosis, Dysporia bronchoenteropancreatica congenita familiaris) is a systemic chronic disease with progressive progression. Which reduces the lives of patients. It is the most common in the population of Caucasian genetic disease, inherited in an autosomal recessive manner. The disease is characterised by clinically chronic obstructive lesions, recurrent respiratory tract infections, Digestive processes and their consequences [7].

Gastrointestinal disorders are the cause of many problems in the care of the child with CF. They are one of the main causes of child malnutrition, reflux disease and shortfall Vitamins (A, D, E, K) [1, 6, 7]. Disorders of pancreatic secretions are the cause of lack of weight gain, short-bodily, fatty, loose, abundant, stinking stools, abdominal pain and bloating [12]. An important role in the care and minimisation of the consequences of these disorders is played by nursing staff, who are required to prepare the child's parents/guardians to implement and comply with the modified nutrition rules since the diagnosis of the disease [5]. Parents Caregivers must be aware that the degree of nutrition affects the general condition of the child, the course of bronchophysical disease, the incidence of exacerbations, psychomotor development and physical performance, and consequently the length and quality of life.

In a child with CF The energy demand is higher than that of its healthy peers and is 130-150% of their daily demand. When assessing the energy needs of a child, consideration should be given to factors such as Child's age and gender, general condition, active. Physical activity of the child, periods of growth and puberty, concomitant diseases (e.g. diabetes), respiratory function and extent of absorption and digestion of nutrients [11].

Tab. 1 Energy demand in patients z CF [11]

Components	**Demand**
Carbohydrates	Ok. 45-50% of energy
Fats	Ok. 35-40 %
Protein	ok. 20 %

Source: Walkowiak J., Lisowska A. Mukowiscydoza (CF). Postępowanie żywieniowe. w: Żywienie dzieci w zdrowiu i chorobie. Krawczyński M. (red.) Wydanie I. HELP MED., Kraków 2008

The way the child is eating with CF depends on his age. Baby The most valuable food is breast milk. Female food contains all enzymes and nutrients, as well as antibodies that protect the child against pathogenic microorganisms. In CF infants who are only breastfeeding a common pro-are too low increments weight. In such cases, it is advisable to feed the baby with high-calorie milk for the initial or next feeding - Milupa Cystilac. In unbreastfed infants, milk is often the primary Food, and in the child an elderly supplement to the diet [15]. Milupa Cystilac In its composition contains the Fats MCT, Increased amount of fat-soluble vitamins,

1 ml of milk delivers 1 kcal [7]. W First year of life of a child's solid foods is introduced according to the scheme as in healthy peers, i.e. Between 4 and 6 months of age. The child's diet should be extended gradually, and each. The new product is introduced individually every 2-3 days by observing the reaction of the child to food consumed [15]. Under the child's nutrition with CF is a high-energy, highly fat-rich diet that is highly saline. In the diet of a child with CF it is necessary to include products from all food groups. In addition, the increase in calorific Food is recommended to add to them: butter, mayonon, cream, yellow cheese. In children with diabetes, it is necessary to modify the number of carbohydrates consumed.

The child's diet with CF should be taken into account: fatty sea fish (mackerel, salmon) and vegetable oils (e.g. linseed oil, rapeseed). The Polish Association of Cystic Fibrosis (PTM) recommends DHA acid supplementation at a dose of 20 – 40 mg/kg/day for children and 1 – 2 G/day for adolescents and adults [5].

An important element of the child's diet with CF is dietary fiber, the absence or too small supply (< 10 g/day) is the cause of constipation and abdominal pain. In contrast, consuming large amounts of dietary fibre causes feelings of fullness and unwillingness to consume meals. Fiber absorbs water softer. While the stool increases its volume, it accelerates the intestinal peristalsis by regulating bowel movements. Insoluble fibre fractions eliminate residual nutrients from the body [4].

Impaired absorption, chronic infections, and inflammation are the main The causes of iron deficiency in children with CF. The reason for this condition is also insufficient its supply along with the diet. Common symptoms of this element deficiency include weakness, faster fatigue, pale skin coatings, more frequent infections, as well as worsening Lung function. In children with CF, systematic monitoring of iron levels increased intake of iron-rich products (e.g. red meat, peas, beans, broccoli, red and green vegetables, egg yolk, whole grain rye bread) is recommended. Small DZ The iron is given as a drop [8].

Probiotics have a major influence on the state of the child's digestive system: they reduce the time of flooding in the gut, restore the natural intestinal microflora, increase the overall immunity of the organism, take part In the production of B vitamins [8].

In order to improve Nutritional status of the child, absorption Proteins and fats are necessary Supplementation with pancreatic enzyme preparations from the first months of life. Pancreatic enzyme preparations taken by the child at appropriate doses contribute to normalization of bowel movements (without fat), minimize gastrointestinal symptoms (flatulence, abdominal pain) and obtain due weight gain. Pancreatic enzyme preparations in the form of acid-resistant, enteral microgranules. To bring the desired effects should be taken at the beginning and during the meal. In the case of infants, the enzyme preparations of the pancreas are given to both babies fed with breast milk as and artificially. The granules are spilling into a teaspoon of milk with the addition of 1 drop of cebione (up to 3-4 months of age). For children from the fifth month of life, enzymes are given with the addition of mousse or apple juice until the child learns to swallow whole capsules. The capsules should not be crushed, poured into milk, soup or directly into the baby's mouth.

Pancreatic enzyme preparations are not given for non-fatty, low-protein, vegetable, fruit (except avocados), chewing gum, lollipos, honey, corn chrublas, juices, coffee, tea,

water. The dose of the preparation is determined individually for each child. The optimal dose of pancreatic enzymes is assumed to be the one that eliminates fat loss in the faeces, normalizes weight gain, causes bad flatness and abdominal pain. Supplementation of pancreatic enzyme preparations increases the risk of developing fibrotic colonopathy. To minimize this risk the maximum dose of lipase/kg bw/day must not exceed 10 thousand. J. [2, 5]. During administration of pancreatic enzyme preparations should be systematically carried out:

-Measurements of the child's weight and their application to the centimetre grid,

-An evaluation of the elastase-1 concentration in the stool (elastase-1 < 100 mcg/G stool is an indication for dose modification preparation),

-The evaluation of the excretion of fats in the stool (excretion of > 7 g/day in persons > 10 years and > 4 – 5 G in children aged 2 – 10 is an indication for dose modification) [5].

In order to facilitate the determination of the optimal dose of enzymes. The nutritional quantity and nutritional value of a patient with CF and/or his carers should be made aware of the need to keep a food diary. In the diary it is necessary to record everything that the child eats during the day, using the household measures: spoon, Cup. The quantity and quality of faeces, vitamins and dietary supplements should also be noted [12].

A patient with a CF problem is the loss of large amounts of water and NaCl through the skin. This process intensisals during the heat, physical activity, Fever, diarrhoea and vomiting. Increased fluid intake is necessary for the proper discharge of secretions and its evacuation outward. Increased risk of hyponatremia necessitates the need for supplementation with chlorinated sod. In the form of tablets or table salt in a way in. Infants and toddlers are given 10% intravenous NaCl solution-1ml = 100 mg NaCl [7].

Tab. 2 Recommended supply of NaCl in mg/kg bw/day [7]

Age of the child	**Recommended supply of NaCl**
Infants	100
1-5 years old	600
6-10 years old	1200
> 11 years old	1800

Source: Mazurczak T. (red). Mukowiscydoza. Instytut matki I dziecka, Warszawa 2006

For older children, adolescents and adults, it is recommended to: repaint dishes, consume NaCl-rich products (e.g. salted meats, Chips, salt sticks, yellow cheese, salted peanuts) and drinking fluids enriched with sodium (isotonic drinks containing electrolytes) [5]

Deficiencies of vitamins, mainly fat-soluble (A, D, E, K) are an indication to monitor their concentration not Less than once a year, and in the case of dose modifications after 3 to 6 months. The correct values for each vitamin are:

-For vitamin A 200 – 800 mcg/L,

-For β-carotene 0.4 – 3.0 mmol/L,

-For vitamin D > 30 ng/ml,

-For vitamin K < 2 ng/ML,

-For vitamin E > 5.4 mg/g.

Low their levels are an indication for their supplementation.

A small proportion of CF patients also have shortages of water-soluble vitamins, the supplementation of which is only indicated if no Supply with a diet. In patients after a small intestine resection, vitamin B12 supplementation is recommended once a month at a dose of 100 mcg [2].

The child with CF irrespective of age and clinical form of the disease Must be under constant control Dietitian. During each visit to a specialised centre, an assessment should be made of Nutritional status of the child and analyse the dietary diary. During the first months of diagnosis, weight measurements should be made at least once per month. If weight gains are unsatisfactory, the weight should be assessed once a week. The decision to increase the calorie intake of meals is taken individually to the patient.

In children who, despite the use of the diet and the intake of pancreatic preparations, do not have satisfactory weight gain, weight loss and gastrointestinal problems are considered for enteral nutrition and outside the gut [5]. In part Children with CF is assumed to be a gastrostomy (direct access to the stomach through the abdominal shell). The nutritional gastrostomy is assumed endoscopic (percutaneous endoscopic PEG gastrostomy), less frequently operatively or laparoscopic. In case of Children up to 16 years of age consent to the establishment of a gastrostomy are signed by the parents/guardians of the child. When the child is 16 years old, the child and his/her parents/guardians sign the consent. To make this form of nutrition safe for your child It is necessary to educate the child and his caregivers about the gastrostomia. This process should be initiated when a decision is taken on the necessity of establishing a gastrostomia. A child with a gastrostomy is accompanied by a clinic leading home enteral nutrition. The clinic nurse teaches the patient/his caregivers:

1. Observation of the skin around the gastrostomia and its proper care

2. Rules for preparing meals and fluids and the ability to administer them

3. Prevent the drain from falling out and stand alone its assumptions in if the actuator is ejected/dropped

4. Maintenance of the Passer

5. Permitted and P/indicated forms of physical activity

The child is given a mixture of their own (mixed) and/or industrial diets with a gastrostomia. Own mixtures should be prepared on the basis of natural or powdered milk, cream, butter, vegetable oils, and starch. Mixtures should not be prepared on the basis of red meat, heavy-weight, high-pitting foods (cabbage, peas, beans(a) and bananas (they operate in a breathtaking). Mixtures should be prepared immediately prior to administration and should have room temperature.

A child with a gastrostomy should also take the enzyme preparations of the pancreas, at a dose determined by the physician. The first dose should be taken before starting the infusion, the second half of the infusion. In some patients, nocturnal nutrition is used (1 ml of infusion should be 1 kcal). With a good tolerance to this form of nutrition is recommended a hypercalcical diet - 1.5 kcal/ml. Homemade Enteral Nutrition from clinical indications is financed by the NFZ. Patients/their caregiers receive a diet contracted by the physician and needed for this equipment [3, 13, 14].

Adherence by the patient and in the case of young children also to their caregivers of the nutrition rules allows providing a suitable calorie and balanced diet, maintaining the correct state of recovery. Vitamins and minerals, which guarantees proper development of the child, extends and improves the quality of life. The nutritional status of the child also affects the course of bronchopulmonary disease and the frequency of its exacerbations.

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